Diseases of the Aorta

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Diseases of the Aorta

- Congenital anomalies of aorta
- Aortic aneurysm (etiology.thorasic.abdominal)
- Aortic dissection(manifestation.treatment)
- Chronic atherosclerotic occlusive disease
- Acute aortic occlusion
- Aortitis

(takayasu. giant cell arteritis. rheumatic.idiopathic.infective aortitis)

Introduction

In adults, AO, diameter is approximately:

- 3 cm at the origin and in the ascending portion,
- 2.5 cm in the descending portion in the thorax,
- and 1.8–2 cm in the abdomen.

The aortic wall consists of:

- a thin intima composed of endothelium, subendothelial connective tissue, and an internal elastic lamina;
- a thick tunica media composed of smooth muscle cells and extracellular matrix;
- and an adventitia composed primarily of connective tissue enclosing the vasa vasorum and nervi vascularis.

- In addition to the conduit function of AO, its viscoelastic and compliant properties serve a buffering function.
- The aorta is distended during systole to allow a portion of the stroke volume and elastic energy to be stored, and it recoils during diastole so that blood continues to flow to the periphery.
- Because of its continuous exposure to high pulsatile pressure and shear stress, the aorta is particularly prone to injury and disease resulting from mechanical trauma.
- AO is more prone to rupture than is any other vessel, especially with the development of aneurysmal dilation, since its wall tension, as governed by Laplace's law ,will be increased.

Congenital Anomalies of the Aorta

- Usually involve the aortic arch and its branches.
- Symptoms such as dysphagia, stridor, and cough may occur if an anomaly causes a ring around or otherwise compresses the esophagus or trachea.
- Anomalies associated with symptoms include:

-double aortic arch,

-origin of the right subclavian artery distal to the left subclavian artery,

-and right-sided aortic arch with an aberrant left subclavian artery.

-A Kommerell's diverticulum is an anatomic remnant of a right aortic arch.

- Most congenital anomalies of the aorta do not cause symptoms and are detected during catheter-based procedures.
- The diagnosis of suspected congenital anomalies of the aorta typically is confirmed by computed tomographic (CT) or magnetic resonance (MR) angiography.
- Surgery is used to treat symptomatic anomalies.

Aortic Aneurysm

- An *aneurysm* is defined as a pathologic dilation of a segment of a blood vessel.
- A *true aneurysm* involves all three layers of the vessel wall and is distinguished from a *pseudoaneurysm*, in which the intimal and medial layers are disrupted and the dilated segment of the aorta is lined by adventitia only and, at times, by perivascular clot.

- May be classified according to gross appearance:
- A *fusiform aneurysm* affects the entire circumference of a segment of the vessel, resulting in a diffusely dilated artery.
- A *saccular aneurysm* In contrast, involves only a portion of the circumference, resulting in an outpouching of the vessel wall.
- Also are classified according to location, i.e., abdominal versus thoracic.
- Aneurysms of the descending thoracic aorta are usually contiguous with infradiaphragmatic aneurysms and are referred to as *thoracoabdominal aortic aneurysms*.

Etiology

- Conditions that cause degradation or abnormal production of the structural components of the aortic wall: elastin and collagen.
- The causes of aortic aneurysms may be broadly categorized as:

-degenerative diseases,

-inherited or developmental diseases,

-infections,

-vasculitis,

-and trauma*

- Inflammation, proteolysis, and biomechanical wall stress contribute to the degenerative processes that characterize most aneurysms of the abdominal and descending thoracic aorta.
- These are mediated by B cell and T cell lymphocytes, macrophages, inflammatory cytokines, and matrix metalloproteinases that degrade elastin and collagen and alter the tensile strength and ability of the aorta to accommodate pulsatile stretch.
- The associated histopathology demonstrates destruction of elastin and collagen, decreased vascular smooth muscle, in-growth of new blood vessels, and inflammation.
- Factors associated with degenerative aortic aneurysms include aging, cigarette smoking, hypercholesterolemia, male sex, and a family history of aortic aneurysms.

Diseases of the Aorta:* Etiology and Associated Factors

- Aortic aneurysm

-Degenerative/atherosclerosis

- Aging
- Cigarette smoking
- Male gender
- Family history

-Cystic medial necrosis

- Marfan syndrome
- Loeys-Dietz syndrome
- Ehlers-Danlos syndrome type IV
- Familial
- Bicuspid aortic valve

-Chronic aortic dissection

-Infective

-Trauma

- Acute aortic syndromes

(aortic dissection, acute intramural hematoma, penetrating atherosclerotic ulcer)

-Atherosclerosis

-Cystic medial necrosis

-Hypertension

-Vasculitis

-Pregnancy

-Trauma

- Aortic occlusion

-Atherosclerosis

-Thromboembolism

- Aortitis
- -Vasculitis

-Takayasu's arteritis

-Giant cell arteritis

- -Rheumatic

-HLA-B27–associated spondyloarthropathies

-Behçet's syndrome

-Cogan's syndrome

- -Idiopathic aortitis
- -Infective

-Syphilis

-Tuberculosis

- -Mycotic (Salmonella, staphylococcal, streptococcal, fungal)

- The most common pathologic condition associated with degenerative aortic aneurysms is *atherosclerosis*.
- Many patients with aortic aneurysms have coexisting risk factors for atherosclerosis , as well as atherosclerosis in other blood vessels.

- *Cystic medial necrosis* is the histopathologic term used to describe the degeneration of collagen and elastic fibers in the tunica media of the aorta as well as the loss of medial cells that are replaced by multiple clefts of mucoid material.

- Cystic medial necrosis characteristically affects the proximal aorta, results in circumferential weakness and dilation, and leads to the development of fusiform aneurysms involving the ascending aorta and the sinuses of Valsalva.
- Is particularly prevalent in patients with Marfan syndrome, Loeys-Dietz syndrome, Ehlers-Danlos syndrome type IV , hypertension, congenital bicuspid aortic valves, and familial thoracic aortic aneurysm syndromes;
- sometimes it appears as an isolated condition in patients without any other apparent disease.

- Familial clusterings of aortic aneurysms occur in 20% of patients, suggesting a hereditary basis for the disease.
- Mutations of the gene that encodes fibrillin-1 are present in patients with Marfan syndrome. Fibrillin-1 is an important component of extracellular micofibrils, which support the architecture of elastic fibers and other connective tissue.
- Deficiency of fibrillin-1 in the extracellular matrix leads to excessive signaling by transforming growth factor (TGF-).

- Loeys-Dietz syndrome is caused by mutations in the genes that encode TGF- receptors 1 (*TGFBR1*) and 2 (*TGFBR2*). Increased signaling by TGF- and mutations of *TGFBR1* and *TGFBR2* may cause thoracic aortic aneurysms.
- Mutations of type III procollagen have been implicated in Ehlers-Danlos type IV syndrome.
- Linkage analysis has identified loci on chromosomes 5q13–14, 11q23.3–q24, and 3p24–25 in several families, although the specific alleles have not been described.

- The infectious causes of aortic aneurysms include syphilis, tuberculosis, and other bacterial infections.
- *Syphilis* is a relatively uncommon cause of aortic aneurysm.
- Syphilitic periaortitis and mesoaortitis damage elastic fibers, resulting in thickening and weakening of the aortic wall.
- Approximately 90% of syphilitic aneurysms are located in the ascending aorta or aortic arch.

- *Tuberculous aneurysms* typically affect the thoracic aorta and result from direct extension of infection from hilar lymph nodes or contiguous abscesses as well as from bacterial seeding.
- Loss of aortic wall elasticity results from granulomatous destruction of the medial layer.

- A *mycotic aneurysm* is a rare condition that develops as a result of staphylococcal, streptococcal, *Salmonella*, or other bacterial or fungal infections of the aorta, usually at an atherosclerotic plaque.
- These aneurysms are usually saccular.
- Blood cultures are often positive and reveal the nature of the infective agent.

- Vasculitides associated with aortic aneurysm include Takayasu's arteritis and giant cell arteritis, which may cause aneurysms of the aortic arch and descending thoracic aorta.
- Spondyloarthropathies such as ankylosing spondylitis, rheumatoid arthritis, psoriatic arthritis, relapsing polychondritis, and reactive arthritis (formerly known as Reiter's syndrome) are associated with dilation of the ascending aorta.

- Aortic aneurysms occur in patients with Behçet's syndrome and Cogan's syndrome.
- Aortic aneurysms also result from idiopathic aortitis.
- *Traumatic aneurysms* may occur after penetrating or nonpenetrating chest trauma and most commonly affect the descending thoracic aorta just beyond the site of insertion of the ligamentum arteriosum.
- Chronic aortic dissections are associated with weakening of the aortic wall that may lead to the development of aneurysmal dilatation.

Thoracic Aortic Aneurysms

- The clinical manifestations and natural history of thoracic aortic aneurysms depend on their location.
- Cystic medial necrosis is the most common pathology associated with ascending aortic aneurysms, whereas atherosclerosis is the condition most frequently associated with aneurysms of the aortic arch and descending thoracic aorta.
- The average growth rate of thoracic aneurysms is 0.1–0.2 cm per year. Thoracic aortic aneurysms associated with Marfan syndrome or aortic dissection may expand at a greater rate.

- The risk of rupture is related to the size of the aneurysm and presence of symptoms,ranging from:

2–3% per year for thoracic aortic aneurysms <4.0 cm

to 7% per year for those >6 cm in diameter.

- Most thoracic aortic aneurysms are asymptomatic; however, compression or erosion of adjacent tissue may cause symptoms such as chest pain, shortness of breath, cough, hoarseness, and dysphagia.
- Aneurysmal dilation of the ascending aorta may cause CHF as a consequence of AR, and compression of the SVC may produce congestion of the head, neck, and upper extremities.

- A chest x-ray may be the first test that suggests the diagnosis of a thoracic aortic aneurysm*.
- Findings include widening of the mediastinal shadow and displacement or compression of the trachea or left mainstem bronchus.
- Echocardiography, particularly TEE, can be used to assess the proximal ascending aorta and descending thoracic aorta.

A chest x-ray of a patient with a thoracic aortic

aneurysm.

- Contrast-enhanced CT, MRI, and conventional invasive aortography are sensitive and specific tests for assessment of aneurysms of the thoracic aorta and involvement of branch vessels*.
- In asymptomatic patients whose aneurysms are too small to justify surgery, noninvasive testing with either contrast-enhanced CT or MRI should be performed at least every 6–12 months to monitor expansion.

An aortogram demonstrating a large fusiform aneurysm of the descending thoracic aorta.*

Treatment: Thoracic Aortic Aneurysms

- BETA-Adrenergic blockers currently are recommended for patients with thoracic aortic aneurysms, particularly those with Marfan syndrome, who have evidence of aortic root dilatation to reduce the rate of further expansion.
- Additional medical therapy to control HTN.
- Recent preliminary studies indicate that ARB and ACEi will reduce the rate of aortic dilation in patients with Marfan syndrome by blocking TGF-signaling; clinical outcome trials of this treatment approach are in progress.

- Operative repair with placement of a prosthetic graft is indicated:

-in symptomatic thoracic aortic aneurysms,

-Ascending aortic diameter is >5.5–6 cm

-Descending thoracic aortic diameter is >6.5-7 cm,

-an aneurysm that has increased by >1 cm/year.

-In Marfan syndrome or bicuspid aortic valve, ascending thoracic aortic aneurysms >5 cm.

-Endovascular repair is an alternative treatment for some descending thoracic aortic aneurysms.

Abdominal Aortic Aneurysms

- Abdominal aortic aneurysms occur more frequently in males than in females, and the incidence increases with age.
- Abdominal aortic aneurysms 4.0 cm may affect 1–2% of men older than 50 years.
- At least 90% of all abdominal aortic aneurysms >4.0 cm are related to atherosclerotic disease, and most of these aneurysms are below the level of the renal arteries.

- Prognosis is related to both the size of the aneurysm and the severity of coexisting CAD and cerebrovascular disease.
- Risk of rupture increases with size of the aneurysm:

-the 5-year risk for aneurysms <5 cm is 1–2%,

- the 5-year risk for aneurysms >5 cm is 20–40% .

- The formation of mural thrombi within aneurysms may predispose to peripheral embolization.

- Commonly produces no symptoms.
- It usually is detected on routine examination as a palpable, pulsatile, expansile, and nontender mass, or it is an incidental finding observed on an abdominal x-ray or ultrasound study performed for other reasons.
- As abdominal aortic aneurysms expand ,they may become painful.
- Some patients complain of strong pulsations in the abdomen; others experience pain in the chest, lower back, or scrotum.

- Aneurysmal pain is usually a harbinger of rupture and represents a medical emergency.
- Acute rupture occurs without any prior warning, and this complication is always life-threatening.
- Rarely, there is leakage of the aneurysm with severe pain and tenderness.
- Acute pain and hypotension occur with rupture of the aneurysm, which requires an emergency operation.

- Abdominal radiography may demonstrate the calcified outline of the aneurysm;
- about 25% of aneurysms are not calcified and cannot be visualized by x-ray imaging.

- An abdominal ultrasound can delineate the transverse and longitudinal dimensions of an abdominal aortic aneurysm and may detect mural thrombus.

- Abdominal ultrasound is useful for serial documentation of aneurysm size and can be used to screen patients at risk for developing an aortic aneurysm.
- ultrasound screening of men 65–74 years was associated with a risk reduction in aneurysm-related death of 42%.
- Screening by ultrasonography is recommended for men 65–75 years who have ever smoked.

- In addition, siblings or offspring of persons with abdominal aortic aneurysms, as well as individuals with thoracic aortic or peripheral arterial aneurysms, should be considered for screening for abdominal aortic aneurysms.
- CT with contrast and MRI are accurate noninvasive tests to determine the location and size of abdominal aortic aneurysms and to plan endovascular or open surgical repair *.

- Contrast aortography may be used for the evaluation of patients with aneurysms, but the procedure carries a small risk of complications such as bleeding, allergic reactions, and atheroembolism.
- Since the presence of mural thrombi may reduce the luminal size, aortography may underestimate the diameter of an aneurysm.

A computed tomographic angiogram (CTA) depicting a fusiform abdominal aortic aneurysm that has been treated with a bifurcated stent graft.

Treatment:
Abdominal Aortic Aneurysms

- Operative repair of the aneurysm with insertion of a prosthetic graft or endovascular placement of an aortic stent graft is indicated for abdominal aortic aneurysms of any size that are expanding rapidly or are associated with symptoms.

- For asymptomatic aneurysms, abdominal aortic aneurysm repair is indicated if the diameter is >5.5 cm.
- In randomized trials of patients with abdominal aortic aneurysms <5.5 cm, there was no difference in the long-term (5- to 8-year) mortality rate between those followed with ultrasound surveillance and those undergoing elective surgical repair.
- Serial noninvasive follow-up of smaller aneurysms (<5 cm) is an alternative to immediate repair.

- The decision to perform an open surgical operation or endovascular repair is based in part on the vascular anatomy and comorbid conditions.
- Endovascular repair of abdominal aortic aneurysms has a lower short-term morbidity rate but a comparable long-term mortality rate with open surgical reconstruction.
- Long-term surveillance with CT or MR aortography is indicated after endovascular repair to detect leaks and possible aneurysm expansion.

- In surgical candidates, careful preoperative cardiac and general medical evaluations are essential.
- Preexisting CAD, CHF, pulmonary disease, DM, and advanced age add to the risk of surgery.
- β-blockers decrease perioperative cardiovascular morbidity and mortality.

- With careful preoperative cardiac evaluation and postoperative care, the operative mortality rate approximates 1–2%.
- After acute rupture, the mortality rate of emergent operation is 45–50%.
- Endovascular repair with stent placement is an emerging approach but at the current time is associated with a mortality rate of approximately 40%.

Acute Aortic Syndromes

- The four major acute aortic syndromes are

-aortic rupture,

-aortic dissection,

-intramural hematoma,

-and penetrating atherosclerotic ulcer.

Aortic dissection

- Aortic dissection is caused by a circumferential or, less frequently, transverse tear of the intima.
- It often occurs along the right lateral wall of the ascending aorta where the hydraulic shear stress is high.
- Another common site is the descending thoracic aorta just below the ligamentum arteriosum.

- The initiating event is either a primary intimal tear with secondary dissection into the media or a medial hemorrhage that dissects into and disrupts the intima.
- The pulsatile aortic flow then dissects along the elastic lamellar plates of the aorta and creates a false lumen.
- The dissection usually propagates distally down the descending aorta and into its major branches, but it may propagate proximally.
- Distal propagation may be limited by atherosclerotic plaque.
- In some cases, a secondary distal intimal disruption occurs, resulting in the reentry of blood from the false to the true lumen.

- There are at least two important pathologic and radiologic variants of aortic dissection:

1-intramural hematoma without an intimal flap

2-and penetrating atherosclerotic ulcer.

- 1-Acute intramural hematoma is thought to result from rupture of the vasa vasorum with hemorrhage into the wall of the aorta.
- Most of these hematomas occur in the descending thoracic aorta.
- Acute intramural hematomas may progress to dissection and rupture.

- 2-Penetrating atherosclerotic ulcers are caused by erosion of a plaque into the aortic media, are usually localized, and are not associated with extensive propagation.
- They are found primarily in the middle and distal portions of the descending thoracic aorta and are associated with extensive atherosclerotic disease.
- The ulcer can erode beyond the internal elastic lamina, leading to medial hematoma, and may progress to false aneurysm formation or rupture.

- Several classification schemes have been developed for thoracic aortic dissections.
- DeBakey and colleagues initially classified aortic dissections as

-type I, in which an intimal tear occurs in the ascending aorta but involves the descending aorta as well;

-type II, in which the dissection is limited to the ascending aorta;

-and type III, in which the intimal tear is located in the descending aorta with distal propagation of the dissection*.

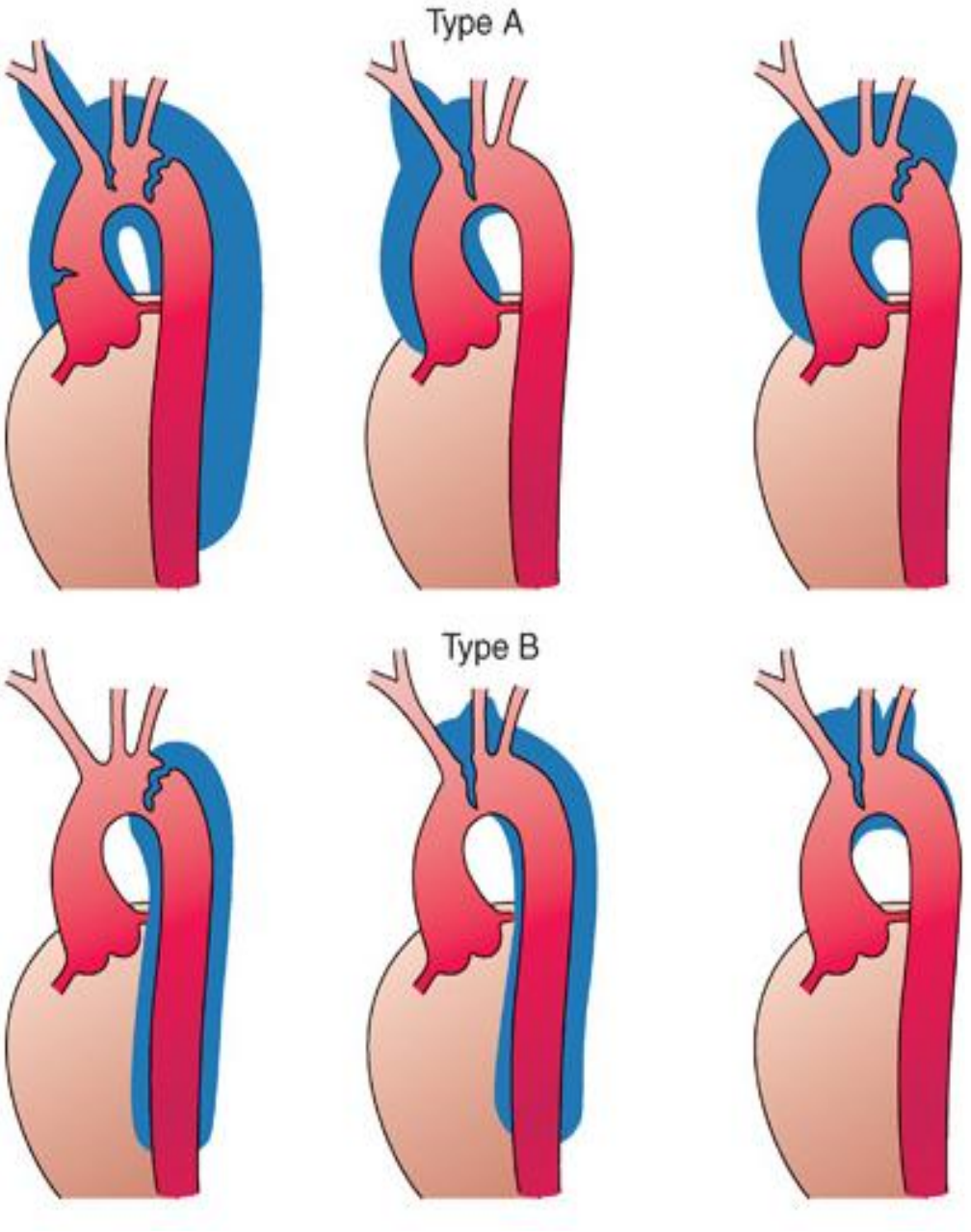
- Another classification (Stanford) is that of

-type A, in which the dissection involves the ascending aorta (proximal dissection),

-type B, in which it is limited to the descending aorta (distal dissection).

From a management standpoint, classification of aortic dissections and intramural hematomas into type A or B is more practical and useful, since DeBakey types I and II are managed in a similar manner.

Figure 248-4**Classification of aortic dissections.**
Stanford classification:
Type A dissections (*top panels*) involve the ascending aorta independent of site of tear and distal extension;
type B dissections (*bottom panels*) involve transverse and/or descending aorta without involvement of the ascending aorta.
DeBakey classification:
Type I dissection involves ascending to descending aorta (*top left*);
type II dissection is limited to ascending or transverse aorta, without descending aorta (*top center* + *top right*);
type III dissection involves descending aorta only (*bottom left*).



- The factors that predispose to aortic dissection include systemic hypertension, a coexisting condition in 70% of patients, and cystic medial necrosis.
- Aortic dissection is the major cause of morbidity and mortality in patients with Marfan syndrome and similarly may affect patients with Ehlers-Danlos syndrome.
- The incidence also is increased in patients with inflammatory aortitis (i.e., Takayasu's arteritis, giant cell arteritis), congenital aortic valve anomalies (e.g., bicuspid valve), coarctation of the aorta, and a history of aortic trauma.
- The risk of dissection is increased in otherwise normal women during third trimester of pregnancy.

Clinical Manifestations

- The peak incidence of aortic dissection is in the sixth and seventh decades.
- Men are more affected than women, 2:1 ratio.
- The presentations of aortic dissection and its variants are the consequences of intimal tear, dissecting hematoma, occlusion of involved arteries, and compression of adjacent tissues.

- Acute aortic dissection presents with the sudden onset of pain, which often is described as very severe and tearing and is associated with diaphoresis.
- The pain may be localized to the front or back of the chest, often the interscapular region, and typically migrates with propagation of the dissection.
- Other symptoms include syncope, dyspnea, and weakness.

- **Physical findings** may include hypertension or hypotension, loss of pulses, aortic regurgitation, pulmonary edema, and neurologic findings due to carotid artery obstruction (hemiplegia, hemianesthesia) or spinal cord ischemia (paraplegia).
- Bowel ischemia, hematuria, and myocardial ischemia have all been observed.

- These clinical manifestations reflect complications resulting from the dissection occluding the major arteries.
- Manifestations may result from the compression of adjacent structures (e.g., superior cervical ganglia, superior vena cava, bronchus, esophagus) by the expanding dissection, causing aneurysmal dilation, and include Horner's syndrome, SVC syndrome, hoarseness, dysphagia, and airway compromise.
- Hemopericardium and cardiac tamponade may complicate a type A lesion with retrograde dissection.

- Acute AR is an important and common (>50%) complication of proximal dissection.
- It is the outcome of either a circumferential tear that widens the aortic root or a disruption of the annulus by a dissecting hematoma that tears a leaflet(s) or displaces it, inferior to the line of closure.
- Signs of AR include bounding pulses, a wide PP, a diastolic murmur often radiating along the right sternal border, and evidence of CHF.
- The clinical manifestations depend on the severity of the regurgitation.

- In dissections involving the ascending aorta, the chest x-ray often reveals a widened superior mediastinum.
- A pleural effusion (usually left-sided) also may be present. This effusion is typically serosanguineous and not indicative of rupture unless accompanied by hypotension and falling hematocrit.
- In dissections of the descending thoracic aorta, a widened mediastinum may be observed on chest x-ray. In addition, the descending aorta may appear to be wider than the ascending portion.

- An ECG that shows no evidence of myocardial ischemia is helpful in distinguishing aortic dissection from MI.
- Rarely, the dissection involves the right or, less commonly, left coronary ostium and causes acute MI.

- The diagnosis of aortic dissection can be established by echocardiography, CT, and MRI.
- Aortography is used less commonly.
- TTE can be performed simply and rapidly and has an overall sensitivity of 60–85% for aortic dissection.
- For diagnosing proximal ascending aortic dissections, its sensitivity exceeds 80%; it is less useful for detecting dissection of the arch and descending thoracic aorta.
- TEE requires greater skill and patient cooperation but is very accurate in identifying dissections of the ascending and descending thoracic aorta but not the arch, achieving 98% sensitivity and approximately 90% specificity.
- Echo also provides important information regarding the presence and severity of AR and pericardial effusion.

- CT and MRI are both highly accurate in identifying the intimal flap and the extent of the dissection and involvement of major arteries; each has a sensitivity and specificity >90%.
- They are useful in recognizing intramural hemorrhage and penetrating ulcers.
- MRI also can detect blood flow, which may be useful in characterizing antegrade versus retrograde dissection.
- The relative utility of TEE, CT, and MRI depends on the availability and expertise in individual institutions as well as on the hemodynamic stability; CT-MRI obviously less suitable for unstable patients.

Treatment: Aortic Dissection

- Medical therapy should be initiated as soon as the diagnosis is considered.
- The patient should be admitted to an ICU for hemodynamic monitoring.
- Unless hypotension is present, therapy should be aimed at reducing cardiac contractility and systemic BP, and thus shear stress.

- For acute dissection, unless contraindicated, β-adrenergic blockers should be administered parenterally, using intravenous propranolol, metoprolol, or the short-acting esmolol to achieve a HR of approximately 60 beats/min.
- +sodium nitroprusside infusion to lower systolic BP to ≤120 mmHg.
- Labetalol, a drug with both α- and β-adrenergic blocking properties, also may be used as a parenteral agent in acute therapy for dissection.

- The calcium channel antagonists verapamil and diltiazem may be used intravenously if nitroprusside or -adrenergic blockers cannot be employed.
- The addition of a parenteral ACE inhibitor such as enalaprilat to a β-adrenergic blocker also may be considered.
- Isolated use of a direct vasodilator such as hydralazine is contraindicated because these agents can increase hydraulic shear and may propagate the dissection.

- Emergent or urgent surgical correction is the preferred treatment for acute ascending aortic dissections and intramural hematomas (type A) and for complicated type B dissections, including those characterized by propagation, compromise of major aortic branches, impending rupture, or continued pain.
- Surgery involves excision of the intimal flap, obliteration of the false lumen, and placement of an interposition graft.
- A composite valve-graft conduit is used if the aortic valve is disrupted.

- The overall in-hospital mortality rate after surgical treatment of patients with aortic dissection is reported to be 15–25%.
- The major causes of perioperative mortality and morbidity include MI, paraplegia, renal failure, tamponade, hemorrhage, and sepsis.
- Endoluminal stent grafts may be considered in selected patients.
- Other transcatheter techniques, such as fenestration of the intimal flaps and stenting of narrowed branch vessels to increase flow to compromised organs, are used in selected patients.

- For uncomplicated and stable distal dissections and intramural hematomas (type B), medical therapy is the preferred treatment.
- The in-hospital mortality rate of medically treated patients with type B dissection is 10–20%.
- Long-term therapy for aortic dissection and intramural hematomas (with or without surgery) consists of control of HTN and reduction of cardiac contractility with the use of β-blockers plus other antihypertensive agents, such as ACEi or calcium antagonists.

- Patients with chronic type B dissection and intramural hematomas should be followed on an outpatient basis every 6–12 months with contrast-enhanced CT or MRI to detect propagation or expansion.
- Marfan syndrome are at high risk for postdissection complications.
- The long-term prognosis for treated dissections is generally good with careful follow-up; the 10-year survival rate is 60%.

Chronic Atherosclerotic Occlusive Disease

- Atherosclerosis may affect the thoracic and abdominal aorta.
- Occlusive aortic disease caused by atherosclerosis usually is confined to the distal abdominal aorta below the renal arteries.
- Frequently the disease extends to the iliac arteries.

- Claudication involves the buttocks, thighs, and calves and may be associated with impotence in males (Leriche syndrome).
- The severity of the symptoms depends on the adequacy of collaterals.
- With sufficient collateral blood flow, a complete occlusion of the abdominal aorta may occur without the development of ischemic symptoms.

- The physical findings include the absence of femoral and other distal pulses bilaterally and the detection of an audible bruit over the abdomen (usually at or below the umbilicus) and the common femoral arteries.
- Atrophic skin, loss of hair, and coolness of the lower extremities usually are observed.
- In advanced ischemia, rubor on dependency and pallor on elevation can be seen.

- The diagnosis usually is established by physical examination and noninvasive testing, including leg pressure measurements, Doppler velocity analysis, pulse volume recordings, and duplex ultrasonography.
- The anatomy may be defined by MRI, CT, or conventional aortography, typically performed when one is considering revascularization.
- Catheter-based endovascular or operative treatment is indicated in patients with lifestyle-limiting or debilitating symptoms of claudication and patients with critical limb ischemia.

Acute Aortic Occlusion

- Acute occlusion in the distal abdominal aorta constitutes a medical emergency because it threatens the viability of the lower extremities; it usually results from an occlusive (saddle) embolus that almost always originates from the heart.
- Rarely, acute occlusion may occur as the result of in situ thrombosis in a preexisting severely narrowed segment of the aorta.

- The clinical picture is one of acute ischemia of the lower extremities.
- Severe rest pain, coolness, and pallor of the lower extremities and the absence of distal pulses bilaterally are the usual manifestations.
- Diagnosis should be established rapidly by MRI, CT, or aortography.
- Emergency thrombectomy or revascularization is indicated.

Aortitis

- May be caused by large vessel vasculitides such as Takayasu's arteritis and giant cell arteritis, rheumatic and HLA-B27–associated spondyloarthropathies, Behçet's syndrome, antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitides, Cogan's syndrome, and infections such as syphilis, tuberculosis, and *Salmonella* or may be associated with retroperitoneal fibrosis.
- Aortitis may result in aneurysmal dilation and AR, occlusion of the aorta and its branch vessels, or acute aortic syndromes.

Takayasu's Arteritis

- This inflammatory disease often affects the ascending aorta and aortic arch, causing obstruction of the aorta and its major arteries.
- Also termed *pulseless disease* because of the frequent occlusion of the large arteries originating from the aorta.
- May involve the descending thoracic and abdominal aorta and occlude large branches such as the renal arteries.
- Aortic aneurysms also may occur.

- Most prevalent in young Asian females but does occur in women of other geographic and ethnic origins and also in young men.
- During the acute stage, fever, malaise, weight loss, and other systemic symptoms may be evident. Elevations of the ESR and CRP are common.
- The chronic stages of the disease, which is intermittently active, present with symptoms related to large artery occlusion, such as upper extremity claudication, cerebral ischemia, and syncope.

- Pathology is a panarteritis characterized by mononuclear cells and occasionally giant cells, with marked intimal hyperplasia, medial and adventitial thickening, and, in the chronic form, fibrotic occlusion.
- Process is progressive, there is no definitive therapy.
- Glucocorticoids and immunosuppressive agents have been reported to be effective in some patients during the acute phase.
- Surgical bypass or endovascular intervention of a critically stenotic artery may be necessary.

Giant Cell Arteritis

- This vasculitis occurs in older individuals and affects women more often than men.
- Primarily large and medium-size arteries are affected.
- The pathology is that of focal granulomatous lesions involving the entire arterial wall; it may be associated with polymyalgia rheumatica.
- Obstruction of medium-size arteries (e.g., temporal and ophthalmic arteries) and major branches of the aorta and the development of aortitis and AR are important complications of the disease.
- High-dose glucocorticoid therapy may be effective when given early.

Rheumatic Aortitis

- Rheumatoid arthritis, ankylosing spondylitis, psoriatic arthritis, reactive arthritis (Reiter's syndrome), relapsing polychondritis, and inflammatory bowel disorders may all be associated with aortitis involving the ascending aorta.
- The inflammatory lesions usually involve the ascending aorta and may extend to the sinuses of Valsalva, the mitral valve, and adjacent myocardium.
- The clinical manifestations are aneurysm, AR, and involvement of the cardiac conduction system.

Idiopathic Aortitis

- Idiopathic abdominal aortitis is characterized by adventitial and periaortic inflammation with thickening of the aortic wall.
- It is associated with abdominal aortic aneurysms and idiopathic retroperitoneal fibrosis.
- Affected individuals may present with vague constitutional symptoms, fever, and abdominal pain.
- Retroperitoneal fibrosis can cause ureteral obstruction and hydronephrosis.
- Glucocorticoids and immunosuppressive agents may reduce the inflammation.

Infective Aortitis

- May result from direct invasion of the aortic wall by bacterial pathogens such as *Staphylococcus*, *Streptococcus*, and *Salmonella* or by fungi.
- These bacteria cause aortitis by infecting the aorta at sites of atherosclerotic plaque.
- Bacterial proteases lead to degradation of collagen, and the ensuing destruction of the aortic wall leads to the formation of a saccular aneurysm referred to as a mycotic aneurysm.
- Mycotic aneurysms have a predilection for the suprarenal abdominal aorta.

- The pathologic characteristics of the aortic wall include acute and chronic inflammation, abscesses, hemorrhage, and necrosis.
- Mycotic aneurysms typically affect the elderly and occur in men three times more frequently than in women.
- Patients may present with fever, sepsis, and chest, back, or abdominal pain; there may have been a preceding diarrheal illness.

- Blood cultures are positive in the majority of patients.
- Both CT and MRI are useful to diagnose mycotic aneurysms.
- Treatment includes antibiotic therapy and surgical removal of the affected part of the aorta and revascularization of the lower extremities with grafts placed in uninfected tissue.

- **Syphilitic aortitis** is a late manifestation of luetic infection that usually affects the proximal ascending aorta, particularly the aortic root, resulting in aortic dilation and aneurysm formation.
- Occasionally involve the arch or descending AO.
- The aneurysms may be saccular or fusiform and are usually asymptomatic, but compression of and erosion into adjacent structures may result in symptoms; rupture also may occur.

- Initial lesion is an obliterative endarteritis of the vasa vasorum, especially in adventitia.
- This is an inflammatory response to the invasion of the adventitia by the spirochetes.
- Destruction of the aortic media occurs as the spirochetes spread into this layer, usually via the lymphatics accompanying the vasa vasorum.
- Destruction of collagen and elastic tissues leads to dilation of the aorta, scar formation, and calcification.
- These changes account for the characteristic radiographic appearance of linear calcification of the ascending aorta.

- The disease typically presents as an incidental chest radiographic finding 15–30 years after initial infection.
- Symptoms may result from AR, narrowing of coronary ostia due to syphilitic aortitis, compression of adjacent structures (e.g., esophagus), or rupture.
- Diagnosis is established by a positive serologic test, i.e., rapid plasmin reagin (RPR) or fluorescent treponemal antibody.
- Treatment includes penicillin and surgical excision and repair.

- QUESTION?